

Systemic Lupus Erythematosus

Risk

- Prevalence: 1:2500 in Northern Europeans and 1:500 in African American population.
- Female:male ratio: 9:1.
- Peak age of onset is between 16-40 y; 20% of SLE pts present prior to age 16 y.

Perioperative Risks

- Increased lupus activity is associated with surgery and stress.
- Infections can initiate lupus or cause a relapse.

Worry About

- CV: Htn, CAD, pericarditis, Libman-Sacks endocarditis with mitral insufficiency, myocarditis, pulmonary
- Pulm: Restrictive lung disease with decreased diffusion capacity
- Renal: Lupus nephritis and renal insufficiency
- Endo: Adrenal insufficiency secondary to chronic corticosteroid use

- Heme: Increased risk of thromboembolism in pts with antiphospholipid antibodies or severe nephrotic syndrome; thrombocytopenia and anemia
- Neurologic: Peripheral neuropathy, delirium, and stroke due to thromboembolism
- Neonatal lupus syndrome: Fetal heart block from maternal autoantibodies that cross placenta

Overview

- Autoantibody-mediated tissue damage results in multisystem organ damage.
- Biopsy demonstrates inflammation, deposition of autoantibodies, and complement in skin and kidneys.
- 15-y survival with lupus is 80%.
- Procainamide, hydralazine, quinidine, clonidine, enalapril, isoniazid, or captopril may cause drug-induced lupus variant.
- Mechanism of increased lupus activity with surgery is unclear but may be related to release of antigens into the bloodstream that bind to circulating antinuclear antibodies to form immune complexes.

Etiology

- Abnormal handling and clearance of cellular debris may lead to autoimmunity against nuclear particles with a high prevalence of specific autoantibodies; ANA, anti-dsDNA, anti-Smith, antiphospholipid.
- A genetic contribution (major histocompatibility complex gene) is important in increasing susceptibility to lupus.
- Ultraviolet radiation is the strongest environmental factor linked to lupus exacerbations.

Usual Treatment

- Lack of specific therapy for SLE.
- Most pts should receive antimalarials (hydroxychloroquine or chloroquine).
- NSAIDs for musculoskeletal symptoms and serositis.
- Immunosuppressive medications, including glucocorticoids, azathioprine, cyclophosphamide, methotrexate, and mycophenolate; belimumab; and monoclonal antibody therapy for treatment resistant disease.
- Smoking cessation; smoking is associated with higher disease activity.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Htn, increased CAD, pericarditis, endocarditis, myocarditis, CHF, conduction blocks, pulm Htn	Chest pain Palpitations Dyspnea	Murmur Pericardial friction rub Peripheral edema	ECG ECHO CXR
RESP	Restrictive lung disease, alveolar hemorrhage, pleural effusion, pulm edema	Pleuritic chest pain Hemoptysis Cough Dyspnea	Pleural rub Cyanosis Decreased lung volume Rales, crackles	CXR PFTs ABG
GI	Gastritis/PUD secondary to medications, lupoid hepatitis, SLE vasculitis resulting in colitis, pancreatitis, and bowel ischemia	N/V Abdominal pain Ileus	Hepatomegaly Splenomegaly Jaundice	LFTs PT/PTT/INR
HEME	Thrombocytopenia, leukopenia, anemia, thromboembolism (lupus "anticoagulant" prolongs aPTT in vitro, but pts have prothrombotic tendency)	Bruising Thrombosis	Lymphadenopathy Splenomegaly	CBC/plts PTT aPL antibodies
RENAL	Glomerulonephritis, nephrotic syndrome, renal insufficiency, renal failure	Fever, hematuria Polyuria Oliguria	Costophrenic tenderness Edema	Urinalysis BUN, Cr, TP, albumin Renal US or scan
CNS	Peripheral neuropathy, stroke, psychosis, fatigue, seizures	Numbness Hemiparesis Paranoid states Hyperirritability	Psychosis Nystagmus, ptosis, diplopia Aphasia	EMG/NCS MRI CT scan EEG
MS/ DERM	Vasculitis and ulceration Arthritis, myalgias, myositis, Raynaud phenomenon	Photosensitivity Ecchymosis or purpura Joint pain or immobility	Malar or butterfly rash Perioral ulcerations	X-ray ANA

Key References: Rahman A, Isenberg DA: Systemic lupus erythematosus, *N Engl J Med* 358(9):929-939, 2008; Ben-Menachem E: Systemic lupus erythematosus: a review for anesthesiologists, *Anesth Analg* 111(3):665-676, 2010.

Perioperative Implications

Preoperative Preparation

- Consider hydrocortisone 50-100 mg IV prior to induction if pt is on chronic steroid therapy.
- Antibiotic prophylaxis if valvular disease is present.

Monitoring

- Caution with arterial line in pts with Raynaud phenomenon.
- Consider PA cath for pulm Htn or CHF.
- Consider Foley cath and CVP/PA cath for fluid titration if renal involvement.

Airway

- Occasionally reduced TMJ, ROM, subglottic stenosis, or cricoarytenoid arthritis manifesting as hoarseness, stridor, or airway obstruction; consider fiberoptic intubation.

Preinduction/Induction

- Consider stress dose corticosteroid therapy.

Maintenance

- No specific agents indicated or contraindicated; consider myocardial function.
- Regional acceptable if no coagulopathy.
- Avoid renally excreted drugs and renal toxins if renal insufficiency is present.
- Cyclophosphamide inhibits plasma cholinesterase and may cause prolonged response to succinylcholine.
- Careful pt positioning if peripheral neuropathies or osteoporosis is present.
- Appropriate thromboprophylaxis.

Adjuncts

- Corticosteroids, supplemental O₂, and careful titration of fluids with renal involvement

Extubation/Postoperative Period

- Reassess respiratory, renal, and CV status prior to extubation.

Anticipated Problems/Concerns

- Adrenal insufficiency from chronic steroid suppression
- Postop infections and pulm complications
- Postextubation laryngeal edema or stridor
- CAD, CHF and arrhythmias
- Renal insufficiency and volume status
- CNS dysfunction, seizures, neuropathy
- Thrombocytopenia, anemia, and thromboembolism
- Lupoid hepatitis